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Living With Spina Bifida: A Historical Perspective

On October 11, 1983, “Baby Jane Doe” was born in Port Jefferson, NY, and diagnosed with hydrocephalus and myelomeningocele, the most common and serious form of spina bifida. Doctors advised her parents that she would die without surgery. With surgery, she would live from 2 to 20 years, paralyzed, bedridden, incontinent, and severely “retarded.” Based on this information, the parents declined treatment. Baby Jane survived however, to become a self-fulfilling prophecy; because she was predicted to have brain damage, surgery was withheld, resulting in damaging infections that might have been avoided with early aggressive treatment. Subsequent litigation and media coverage concerning Baby Jane Doe focused on the issue of “quality of life.” (The story of Baby Jane Doe is recounted in many places. See, for example refs 1 and 2.)

Before 1960, the survival rate for all forms of spina bifida was 10% to 12%.³ Surgeons typically postponed treatment until age 2, believing that only the strongest would survive that long.^{4,5} Before the introduction of antibiotics in the 1940s, most infants with myelomeningocele succumbed to meningitis.

Hydrocephalus also contributed to mortality; the development of shunts in the late 1950s thus revolutionized treatment of myelomeningocele.⁶ Those who survived meningitis and hydrocephalus faced renal complications. In the 1960s, procedures for urinary diversion were developed to preserve renal function and allow children to gain social continence.⁷

By the mid-1960s, improvements in treatment produced higher survival rates and lesser degrees of disability. In 1967, W.J.W. Sharrard, Robert Zachary, and John Lorber, pediatric surgeons at Children’s Hospital, Sheffield, UK, reviewed the cases of 526 children born between 1955 and 1962 and treated for myelomeningocele. They concluded that there was “no place for the selection of patients for conservative treatment rather than operative treatment on the grounds of paralysis, deformity or hydrocephalus present at birth.” As a result of this and similar studies published in the mid-1960s, most centers in the United Kingdom and United States adopted the practice of operating within 12 to 48 hours of birth on all infants who did not have other defects incompatible with life.⁸

Zachary became the leading voice of optimism regarding children with myelomeningocele. He recognized that the social environment shaped the experience of disability and that questions about treatment were really questions about the place of people with disabilities in society.⁴ Acknowledging the unrealized potential of children with myelomeningocele, Zachary maintained that “extreme disability is not synonymous with unhappiness and we are only at the beginning of finding ways of developing the capabilities of these patients....”⁹

Higher survival rates came at a price. The shunts that so effectively treated hydrocephalus required frequent revision. The long-term effects of impaired bladder function resulted in renal failure becoming the leading cause of death for individuals with myelomeningocele in the 1970s. Longer survival also meant more orthopedic surgeries to facilitate walking. Children often underwent dozens of surgeries, spending months in hospitals, with psychological and educational consequences.¹⁰

In 1971, Zachary’s coauthor, John Lorber, reversed the position he had taken 4 years earlier. Discouraged by what he

viewed as the poor quality of life of children with myelomeningocele, Lorber argued for abandoning the practice of treating most infants with the defect in favor of treating only the most promising cases. In stark contrast to Zachary, he wrote that the “majority of children” with myelomeningocele “have very few or no friends; most are left without jobs, they have no chance of love or marriage, and when their exhausted parents can no longer cope, they will end their days in an institution.”¹¹ Rejection by the able-bodied world, in Lorber’s view, justified nontreatment. Based on his views about how disabilities translated into an unacceptable quality of life, Lorber established criteria to predict at birth which lives would be “worth saving.”¹² Lorber’s criteria were widely adopted internationally during the 1970s and 1980s, but a vocal minority objected. They argued that implementing the criteria constituted a denial of ordinary care, because Lorber advocated withholding antibiotics and heavily sedating infants so that they would not demand feeding (see, for example, refs 12 and 13). Furthermore, many physicians did not follow those protocols for ensuring death, resulting in untreated survivors who ended up more severely impaired than would have been the case with immediate treatment.

The trans-Atlantic debate about selective nontreatment of myelomeningocele helped shape the emerging profession of bioethics in the 1970s, as pioneer bioethicists addressed quality-of-life issues related to disability.¹⁴ Those bioethical writings, along with Lorber’s criteria, influenced a clinical research study conducted at the University of Oklahoma Health Sciences Center and published in *Pediatrics* in 1983.¹⁵ From 1977 to 1982, investigators tested the effectiveness of a multidisciplinary approach to nontreatment decision-making for infants with myelomeningocele.

A team evaluated 69 infants, recommending 33 for nontreatment; all 24 whose parents concurred died. In addition to predicting “future ambulatory potential” and “prognosis for intellect,” the team maintained that decisions regarding treatment could vary for infants with the same physical and intellectual prognoses, depending on socioeconomic factors. Cuts in government funding for indigent medical care and special education diminished the quality of life for poorer children with disabilities, they argued, thereby justifying the nontreatment and deaths of some infants.¹⁵ Whereas Lorber’s views had been widely embraced, the authors of the Oklahoma study suffered a backlash from the American medical community. The study was challenged on ethical grounds for both its design and execution, although litigation resulted in favorable rulings for the researchers and the university.¹⁶ One critic wrote: “It is the rare child who is so severely impaired that independent living is automatically excluded as a goal. What are exclusive are our own biases toward the physically and mentally impaired, and our own unwillingness to provide a proper, easily accessible educational environment that maximizes performance.”¹⁷

Ironically, the debate over *whether* to treat children with myelomeningocele continued into the 1980s, even as prognoses for such children improved. Clean intermittent catheterization “all but eliminated the mortality rate associated with urological complications.”¹⁸ Computed tomography technology and better shunt design improved treatment of hydrocephalus and demonstrated that treating even severe cases usually resulted in “normal” intelligence. Improvements in neonatal intensive care helped infants recover from complications. Spina bifida clinics emerged where children could receive

continuous coordinated care from multidisciplinary teams. Changing attitudes toward disability, reflected in passage of the Education for All Handicapped Children Act (1975) and the disability rights movement, promised better opportunities for future independent living and employment. By 1984, most children living with myelomeningocele could achieve independent mobility and social continence, whereas most cases of intellectual disability could be prevented by early aggressive treatment. “Technological change has been so rapid and so progressive,” pediatric neurosurgeon Anthony Gallo wrote, “that it is virtually impossible to give a completely accurate prognosis of an infant born with meningomyelocele and hydrocephalus.”¹⁹ Unfortunately, the pace of change meant that many physicians were simply unequipped to help parents, like Baby Jane Doe’s, make informed decisions.¹⁰

The experience of living with myelomeningocele has improved significantly since the 1960s because of developments in medical technology and changing attitudes toward disability. Among medical professionals, however, a strong current of pessimism about life with the condition has persisted. (Medical pessimism regarding disability has been extensively documented. For an example specific to spina bifida see ref 18, and also ref 19.) Having encountered many misinformed parents in their pioneering work in fetal surgery, Bruner and Tulipan in 2004 wrote an editorial urging other physicians to “tell the truth about spina bifida.”²⁰ de Jong²¹ cites that editorial in his critique of the Groningen Protocol, which permits the euthanasia of newborns with myelomeningocele in the Netherlands. According to de Jong,²¹ the Protocol’s application to myelomeningocele is based on outdated and, in some cases, false information about the condition, leading to overly pessimistic assessments in most cases. Newly

diagnosed parents participating in online forums, such as BabyCenter, routinely report being pressured to abort by obstetricians or maternal/fetal medicine specialists who present the “worst case scenario” but call it a “prognosis.” Adults with myelomeningocele participating in the online forum SpinaBifida-Connection report the impact that negative attitudes of others, especially educators and medical professionals, have had on their own lives. Children and adults successfully living with spina bifida are often portrayed as having miraculously overcome the odds. In reality, the odds have been misrepresented in ways that have cost countless born and unborn lives and sometimes negatively shaped the experiences of those who live with spina bifida.

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